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THE VALUE OF TROPHIC BONE CHANGES IN THE DIAGNOSIS OF LEPROSY *

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The diagnosis of anesthetic leprosy is often very difficult, as the bacillus of Hansen is obtained in only a small proportion of the early cases and consequently the diagnosis usually rests on the symptoms and signs of the neuritis and the gross trophic changes arising from it. These trophoneurotic changes are usually most marked in the extremities and may affect any or all of the tissues of the extremity.

The changes which take place in the nerves, muscles, subcutaneous tissue and skin have been emphasized by the writers on this disease and are well known to all. But the corresponding changes occurring in the bone, although of equal if not of greater diagnostic significance, have been simply mentioned in the descriptions of this disease. This is readily understood when we consider the difficulty of determining these changes early in the disease, before it has advanced to such a stage that these lesions become curious and interesting features of the process, rather than aids in diagnosis.

These bone lesions have been described by several writers. In 1886, Heiberg[1] called attention to peculiar atrophies of bone occurring in this disease. In 1898, Miura[2] described the pathological changes and also followed these changes with the aid of the *x*-ray. Hansen and Looft[3] also described the various processes. More recently Hirschberg and Bichler[4] have shown that a true leprous osteomyelitis and periostitis often occurs. In 1905, Deycke[5] traced the sequence of certain of these changes by means of the *x*-ray, and in 1909, Harbitz,[6] from a study of pathological speci-

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* Read at the meeting of the Canal Zone Medical Association, Nov. 9, 1910.

1. Heiberg, H.: Om lepra mutilans og trofoneurotiske forandringer ved spedalskhed, klin. Aarb., 1886, iii.

2. Miura, K.: Ueber die Veränderungen der Knochen an den Händen und Füssen bei Lepra mutilans, Mitt. a. d. med. Fac. d. Kais. jap. Univ. zu Tokio, 1898, iv, 107.

3. Hansen and Looft: Die Lepra vom klinischen und pathologisch-anatomischen Standpunkte, Biblioth. med. int., 1894, ii, 2.

4. Hirschberg and Bichler: Lepra der Knochen, Dermat. Ztschr., 1909, xvi, 415.

5. Deycke: Knochenveränderungen bei Lepra nervorum in Roentgengraph, Fortschr. a. d. Geb. d. Roentgenstrahlen, 1905-6, ix, 9.

6. Harbitz: Trophoneurotic Changes in Bones and Joints in Leprosy, THE ARCHIVES INT. MED., 1910, vi, 147.

mens, called attention to a peculiar atrophy in this disease which he describes as concentric bone atrophy.

The pathological changes in the bones in leprosy occur in three different ways:

1. The true trophic disturbance due to the altered innervation, in which there takes place a simple atrophy or gradual absorption of the bone.

2. An osteomyelitis or periostitis caused by the lepra bacillus.

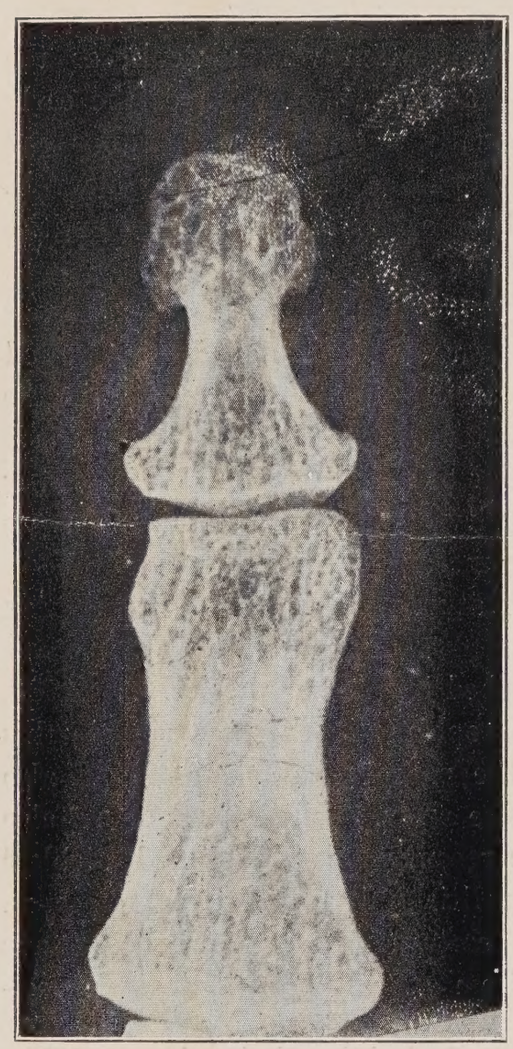

Figure 1.

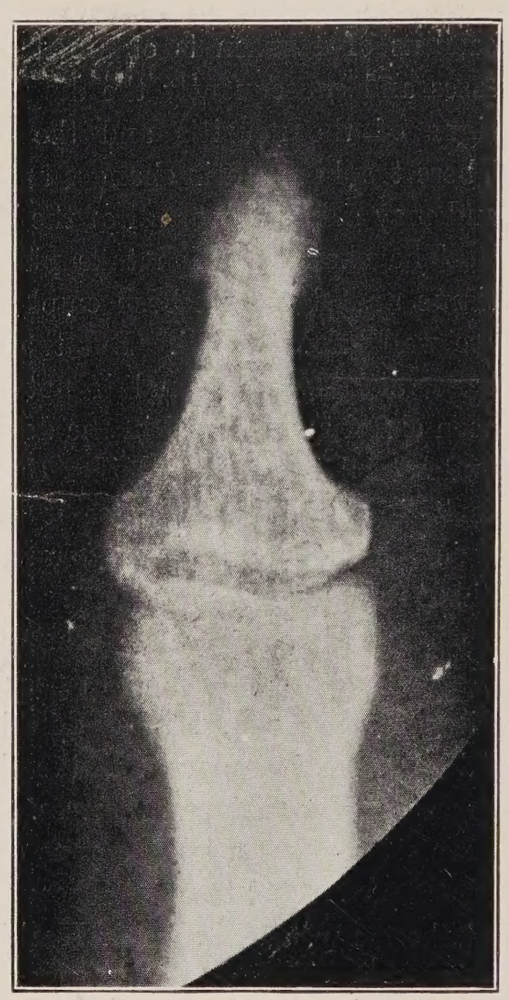

Figure 2.

Fig. 1.—Middle and distal phalanges of a normal index-finger.

Fig. 2.—Beginning absorption of the bulbous tip of the distal phalanx of the thumb, and an associated narrowing of the shaft. Both the shaft and the distal end are involved.

3. A necrosis or inflammation of either of the above arising from secondary involvement by pyogenic organisms.

These processes occur mainly in the fingers and toes and lead to a loss of the digits, from which this form of the disease gets its name of lepra mutilans.

Clinically the process in which a finger or toe is cast off or disappears presents two entirely different forms. The one shows simple atrophy and absorption of the digit without gross disturbance. The other presents an associated inflammatory process of various types. The former would represent a pure trophic type, and the latter a tropho-inflammatory method of the mutilating process.

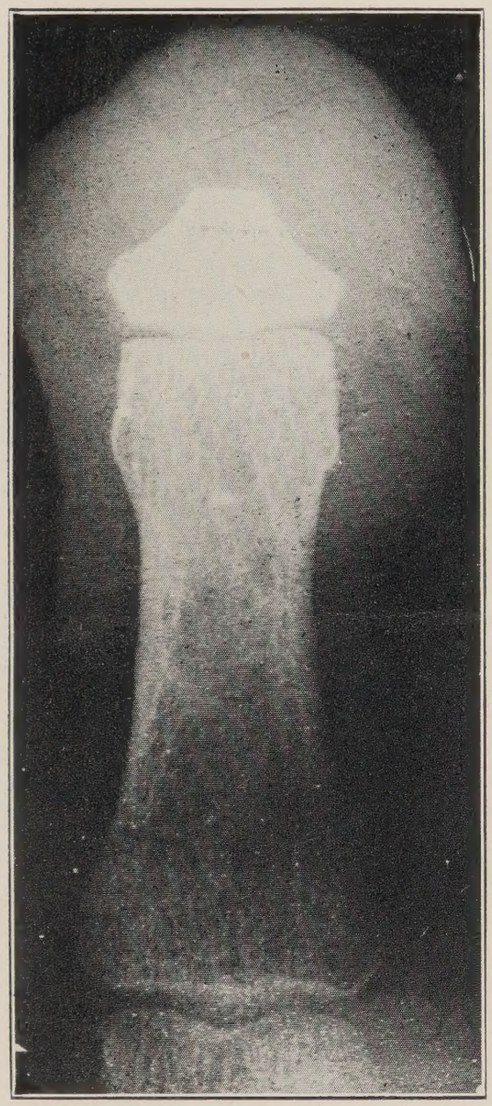

Figure 3.

Figure 4

Fig. 3.—Absorption of the greater part of the bulbous tip of the distal phalanx of the index-finger. No change in the shaft.

Fig. 4.—Absorption and disappearance of the tip and shaft of the distal phalanx of the finger, only the base remaining.

In the tropho-inflammatory type there occurs a casting off of the digits in various ways, but in all the ways there is the underlying trophic change to which is added a secondary infection arising by different methods and thus causing the clinical variations. A perforating ulcer may form, which gradually extends to the shaft of a bone or to a joint

and thus through a chronic suppurative process results in a casting off of the bone or that portion of the member distal to the ulcer. An abscess close to the bone may destroy the periosteum and lead to necrosis of the bone, or a necrosis of the bone of the atrophic type might become secondarily infected and thus lead to loss of the bone with later shrinking of the soft parts. A gangrene-like process might occur through involvement

Figure 5.

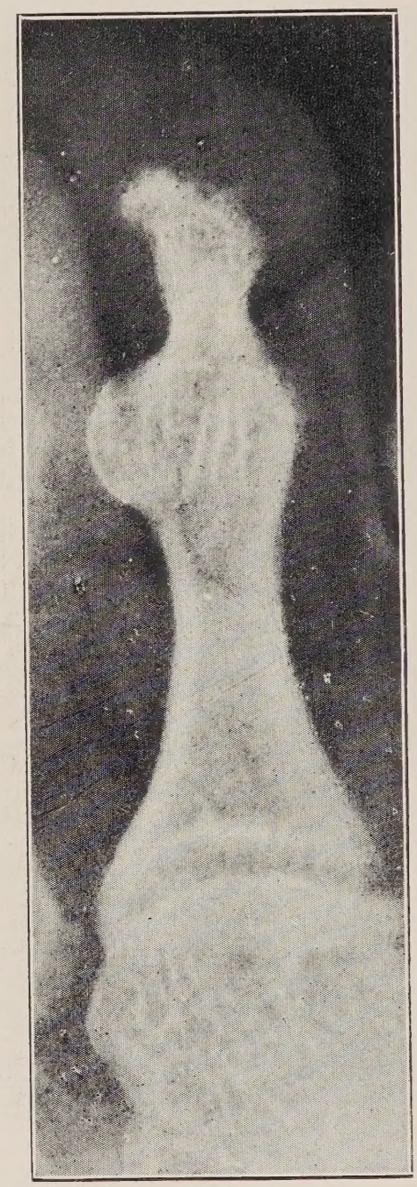

Figure 6.

Fig. 5.—Typical concentric atrophy with absorption and shrinking of the distal phalanx of the middle finger, the process affecting both tip and shaft but more marked in the shaft. It also involves the shaft of the middle phalanx.

Fig. 6.—Typical concentric atrophy of all the phalanges of the third toe; the distal phalanx almost absorbed; the middle phalanx markedly shrunken; the proximal phalanx showing incipient atrophy.

of the vessels by the leprous formations and thus cause the loss of a finger or toe. All of these forms depend on the secondary inflammatory process and consequently show no lesions characteristic of the disease.

The pure trophic type has been variously described. Scheube[7] speaks of it as "simple absorption of the tissues." Manson[8] describes it as "a curious interstitial absorption of one or more phalanges, the shaft of the phalanx wasting more rapidly than either of the articular surfaces." Harbitz speaks of it as "atrophy of the bone of characteristic appearance. The atrophy is concentric and chiefly involves the most peripheral parts of the phalanx."

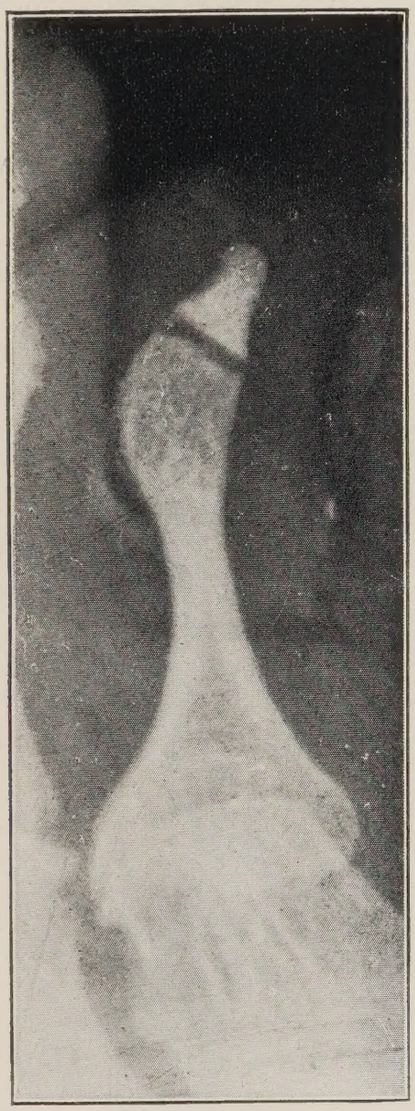

Figure 7.

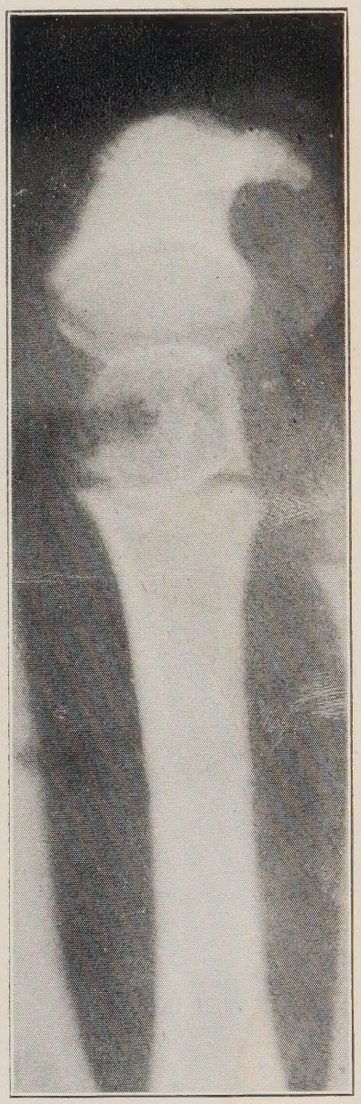

Figure 8.

Fig. 7.—Advanced concentric atrophy of all phalanges of fourth toe. The distal and middle phalanges are almost entirely absorbed. The proximal phalanx shows marked shrinking and narrowing of the shaft.

Fig. 8.—Very advanced concentric atrophy of all the phalanges of the ring finger.

In addition to this simple atrophy of the bone there is described an ainhum-like process which leads to the loss of a toe but which cannot

7. Scheube: Diseases of Warm Countries, 1904, Edition 2, p. 255.
8. Manson: Tropical Diseases, 1909, Edition 4, p. 546.

be distinguished from true ainhum except that other manifestations of leprosy are present.

The relative frequency of these two processes in the causation of mutilation is stated differently by writers. Harbitz says that "many, perhaps the majority, are dependent on ulcerative necrosis and chronic suppurative inflammation." Van Bergman and Guill say that "the

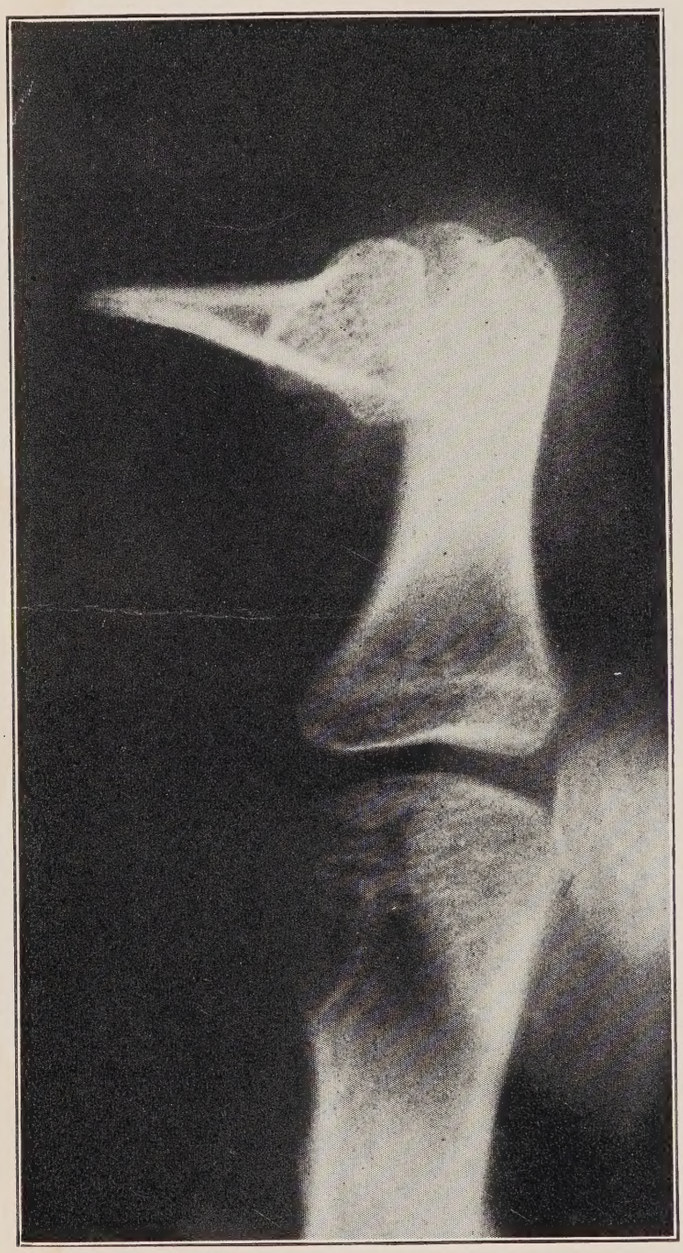

Fig. 9.—E. L., leprosy suspect. Typical concentric atrophy of distal phalanx of right thumb.

phalanges become necrotic and are cast off through suppuration. This is the case as a rule." Balz describes simple absorption of the tissue as the usual cause of the mutilation. From a study of the cases accessible here, I believe that mutilations arise in almost equal numbers by the two meth-

ods, but that if we could examine the case before the secondary infective processes took place, we would find that the great majority of these would show trophic changes in the bone. Most of the mutilating processes, at present in the course of development in the cases here, are taking place by the trophic method. In the course of years a certain number of these will undoubtedly fall into the other class, secondary infections arising in them from trauma or other causes. This is shown in the case of a boy at Palo Seco who has lost all the fingers and thumbs of both hands. Of

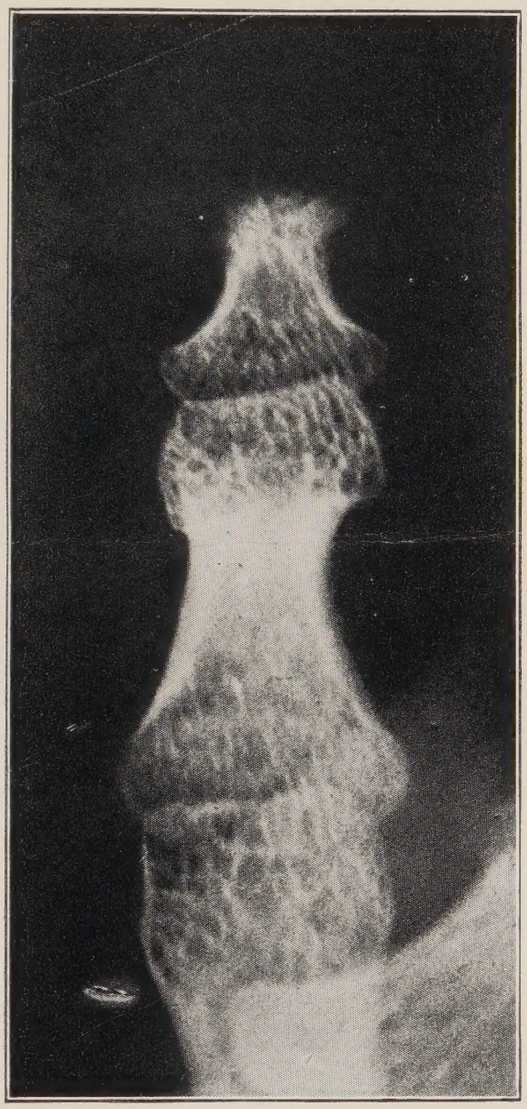

Fig. 10.—E. L., leprosy suspect. Typical distal atrophy of distal phalanx of index-finger of right hand.

the ten digits lost, four were removed on account of suppurative processes, and in the other six there was simply a gradual atrophy and shrinking of the fingers.

The pure trophic type presents bone lesions which are characteristic of the disease. This has been shown by Harbitz in his work on pathological specimens showing characteristic trophic changes late in the dis-

ease. These trophic changes in many cases begin early and present definite pictures which give important additional aid in the diagnosis. This trophic disturbance usually begins in the tip of the distal phalanx and may involve one or more fingers. As the process advances in the distal phalanx, changes are found in the middle and later in the proximal phalanx, and may even extend to the metacarpal or metatarsal bones. This same trophic change extends to other bones, but we are interested only in the earliest changes when the diagnosis of the case might be doubtful. These early changes are easily observed by means of the *x*-ray.

During the past two years we have been giving special attention te the bone changes in the cases of leprosy which have been brought before the medical examining board of this hospital. The study of the *x*-ray plates taken in these cases and also in several others from Palo Seco has revealed bone changes varying greatly in degree. These changes we have arranged according to the character and extent of the bone involvement so as to illustrate the development and progress of the trophic bone lesions of this disease. In order to understand these, especially the earlier lesions, the appearance of the distal phalanx with its cellular bulbous tip must be recalled (Fig. 1).

The earliest change noticed is in the bulbous tip of the terminal phalanx. This becomes smaller and shrinks, generally with some associated narrowing of the shaft (Fig. 2), or the process may affect the bulbous tip mainly, in which case there appears to be a definite absorption starting at the most distal point, and progressing almost transversely toward the base (Fig. 3). As this process progresses, the tip disappears and then the shaft becomes gradually absorbed, and a stage is reached in which only the base of the distal phalanx remains (Fig. 4). So far the process shows a distal type of atrophy in which the absorption is mainly in the distal extremity of the bone. In the true concentric type, when the process becomes more advanced, the distal phalanx shows marked shrinking and shortening in both the shaft and the distal extremity, and there is also a shrinking in the shaft of the middle phalanx as is shown by the increase of the concave outline of its borders (Fig. 5). In this form the process involves the shaft more than the distal extremity. In a later stage the terminal phalanx almost entirely disappears, being represented by a very slight shadow on the *x*-ray plate. The middle phalanx becomes greatly shrunken and smaller, and the proximal phalanx shows the incipient changes in the narrowing and shrinking of the shaft (Fig. 6). The interphalangeal joint spaces disappear, the joints becoming partially or totally ankylosed. In a more advanced stage the distal and middle phalanges are reduced to very small fragments, and the proximal phalanx shows a very marked concentric atrophy, almost obliterating the shaft of the bone, with marked absorption of the distal extremity (Figs. 7 and 8).

The same changes may progress farther and involve the metatarsal and metacarpal bones, but by this time the disease is usually so far advanced that the diagnosis is comparatively easy from the gross signs. These later changes are well shown in the pathological specimens radiographed by Harbitz, and also in some of the cases reported by Deycke.

The successive changes shown in these enlargements have been taken from positive leprosy cases and represent the gradually increasing lesions in the progress of the disease. The earliest changes occur in the distal extremity of the terminal phalanx and on account of its small size in the ordinary *x*-ray plate, it is readily overlooked. Also radiographs from suspected cases usually reveal only one or two of these lesions at the most.

In the usual case of anesthetic leprosy, the simple trophic lesion alone is not always found, for frequently secondary inflammatory changes have taken place, as would be expected when we consider that a perforating ulcer is one of the earliest and most common signs of this form of the disease. Accordingly in many cases there occurs a combination of the concentric bone atrophy with ulcerative bone necrosis. This combination occurs in the majority of the cases of anesthetic leprosy, but even with this additional bone necrosis, the true trophic lesion can be readily made out.

These trophic bone changes may appear early and are of great aid in the diagnosis of the disease, as they furnish an additional sign and one quite characteristic. This is shown in the case of a recent leprosy suspect under examination here. The case was that of a little Jamaican girl, 18 years old, presenting an anesthetic type of the disease. She has marked analgesia of both hands and feet. The hands show the beginning contractures of the claw-hand. In the left hand there is beginning ankylosis of the joints of the little and ring fingers with simple atrophy of the tip of the index-finger. The right hand shows a later stage of the claw-hand involving all four fingers. There is marked atrophy of the tip of the thumb and partial atrophy of the tip of the index-finger. In both feet the second toe is swollen and discolored and presents an ulcer at the tip. There is a neuritis of both peroneal nerves, the child walking with a partial foot-drop.

Examinations of the mucous membrane of the nose and of all lesions were negative for the bacillus of Hansen four months ago, and remain negative at this time.

Radiographs of the hands show typical concentric atrophy of the distal phalanx of the right thumb and of the distal and middle phalanges of the left little finger. They also show typical distal atrophy of the terminal phalanges of the index-fingers; slight distal atrophy of the end phalanx of the left middle finger; and very marked in the right middle finger, the entire distal phalanx having been absorbed. The last repre-

sents the most advanced lesion of distal atrophy we have found in the cases here (Figs. 9 and 10).

In any consideration of the value of these trophic changes it is necessary to determine under what conditions more or less similar bone changes might occur. There are some conditions which produce bone lesions somewhat resembling these, but as a rule they can be readily differentiated by a proper history and examination of the case. Similar changes could arise as a result of previous injuries; in the atrophy of disuse as might occur in paralysis or in nerve lesions; in Morvan's disease, and in ainhum. Also the race and occupation of the individual must be taken into consideration in the interpretation of the radiograph.

In a study of the early lesions occurring in the feet, a knowledge of the race and occupation of the individual becomes necessary, for in the barefoot native the phalanges of the toes are broader, larger, and far better developed than in the members of the races wearing shoes. This is well shown in the radiographs of the feet of Americans when compared with those of the islanders or natives.

In the case of injuries involving the bones of the phalanges changes more or less similar to progressing distal atrophy may occur, but the history of the case would serve to differentiate this condition.

Similar lesions might occur following injuries to nerves or in the atrophy of disuse, but in these cases the lesions would be limited to a certain definite area and all the bones in this area would be similarly involved. Also an extra point of differentiation could be obtained from the history of the case.

In syringomyelia, more especially that variety known as Morvan's disease, a mutilating process takes place which in great part resembles the same process in anesthetic leprosy. The description of this process as detailed in the literature would point to its occurrence by means of an ulcerative-necrotic method rather than by true concentric atrophy, as the descriptions of Morvan's disease speak of this mutilation as occurring through an inflammatory process.

In ainhum there is a peculiar trophic disturbance which shows bone changes very similar to those occurring in leprosy. The process involves the proximal phalanx primarily and there is a lengthening with narrowing and absorption of the shaft rather than a shortening and absorption as occurs in the concentric atrophy of leprosy. But what the bone change would be in a case of leprosy in which the loss of the toe was taking place by an ainhum-like process we are unable to say, as we have seen no such case.

In a study of the radiographs taken from the various cases of leprosy accessible here, there are two main methods by which the atrophy of the bone takes place. The one could be called progressive distal atrophy. It

starts at the distal extremity of the terminal phalanx and progresses almost transversely toward the base. We have not been able to follow this process beyond the distal phalanx. It appears to be the more rapid method of mutilation and occurs more often in cases of a mixed type of the disease. It also bears a close resemblance to the bone changes following injuries, or to amputation stumps. The other method presents the true concentric atrophy well described by Harbitz. In this there is a general shrinking and absorption of the distal portion and shaft of a phalanx simultaneously. This process is very characteristic of the disease, but a similar process might occur in the atrophy of disuse or in a severe injury or lesion of the innervating nerve.

These trophic bone lesions, while not absolutely pathognomonic of the disease, are very characteristic of it. Any condition which might present a similar bone lesion could be readily differentiated by the previous history of injury or disease taken in connection with the examination of the case. Of all the trophic lesions of leprosy these are the most characteristic, and while not occurring in all cases they are found in the majority and very often sufficiently early to be of great aid in the diagnosis. It is in the anesthetic form of leprosy especially that these two forms of bone changes—the progressive distal atrophy and the concentric atrophy—furnish an additional diagnostic sign and one very characteristic of the disease.

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